

*Leading article*

## Long term prognosis of Crohn's disease with onset in childhood and adolescence

The incidence of Crohn's disease rises sharply from the age of 13 and the disease is rare below the age of 9 years. About a quarter of all cases are diagnosed before the age of 21.<sup>1</sup> No epidemiological studies of incidence confined to an artificial 'paediatric' age group have been done, but in the Sick Children's Hospital in Toronto eight new cases of Crohn's disease compared with 12 patients with coeliac disease are seen annually,<sup>2</sup> drawn from a population of about five million.

As paediatric gastroenterology has become established, there has been an increasing interest in the presentation, course, and prognosis of Crohn's disease in childhood. The paper by Puntis *et al* in this issue describes the largest series of paediatric patients collected in the United Kingdom.<sup>3-5</sup> The patients have been followed up for a long period by the same group (on average 16.5 years), so that estimates of prognosis in the long term are more likely to be accurate than in other studies with a short follow up, or a review by questionnaire.<sup>6</sup>

Many studies have shown that the overall prognosis of Crohn's disease in adults is better than formerly supposed, with a cumulative risk of death about twice normal during long term follow up. Cooke and his colleagues calculated a cumulative relative risk of 10 times in the first five years after diagnosis, falling to 2.4-fold at 15 years.<sup>7</sup> This low mortality may disguise considerable morbidity. Follow up of 512 patients suffering from Crohn's disease with onset in childhood showed that only a third regarded themselves in optimal health on average 7.7 years later. In the Birmingham series on the other hand, 52 of the 58 live patients considered themselves well, despite recurrent disease in 14. This is confirmed by the adult series from the same centre where of 121 live adults, only two were not leading normal lives free of restriction.<sup>7</sup> Previous studies, however, have shown that the patients' estimate of their state of health correlates more closely with their personality than with the extent or activity of their disease.<sup>4</sup>

Accurate mortality statistics of Crohn's disease presenting in childhood are impossible to extract from published studies which only quote crude figures. These may not be representative as they come from major referral centres, but nine of the 67 children in the Birmingham series died (average follow up period 16.5 years) while the death rate in the 512 American children studied by Farmer and Mitchener was low at 2.4% (13 children), but the average follow up was only 7.7 years.

It is likely that the overall prognosis of Crohn's disease presenting in childhood is similar to that of the adult disease although there are a number of unique features when the disease starts early in life.

### GROWTH RETARDATION

Not surprisingly, the symptoms of Crohn's disease in childhood which are not dissimilar to those in adults may be subtle, although comfort should be

derived from the fact that at presentation either the haemoglobin, albumin, or acute phase proteins were abnormal in all cases in the Birmingham study.<sup>3</sup> It would be tragic if the concern to exclude a diagnosis of Crohn's disease led to overinvestigation of the 10% of all children who present at some stage during development with abdominal pain. Indeed, O'Donoghue and Dawson<sup>5</sup> found no proof in their 32 children that abdominal pain without other symptoms was an early feature of Crohn's disease.

The unique symptom of Crohn's disease presenting in childhood is growth retardation, and this affects at some stage 30% of the patients in the study reported in this issue.<sup>3</sup> There are many published, but inadequate reports of growth retardation, which do not specify whether this relates to height (more important) or to weight, and take no account of familial factors that determine growth rates. Early reports imply that growth retardation may precede other symptoms of Crohn's disease in childhood by several years, but only four patients studied by Puntis and colleagues<sup>3</sup> presented with weight loss or growth retardation alone; this agrees with other recent analyses.<sup>8</sup> It is now recognised that retardation of linear growth is usually caused by nutritional deficiency and any hormonal changes are secondary to this.<sup>9</sup> Thus accelerated rates of growth can be induced by total parenteral nutrition (TPN),<sup>10</sup> or by supplementation with elemental diets.<sup>11</sup> Unfortunately elemental diets are unpalatable and it is difficult to persuade children to eat them, while a period of TPN seems to be frequently followed by rapid recurrence of the disease. In carefully selected groups of prepubertal patients with Crohn's disease surgery can induce a growth spurt to at least the 10th centile. This reversal of growth retardation was recorded in eight of 12 patients with ileocolic disease and two of four patients with colonic disease in the Birmingham study.<sup>3</sup> These encouraging figures contrast with those of Homer *et al*<sup>12</sup> where only two of 11 prepubertal patients with failure of linear growth showed catch up growth postoperatively. Thirty per cent of patients undergoing operation in Homer's whole series had recurrence of disease within two years and this was always associated with failure of catch up growth. Likewise children with incomplete excision at the time of initial operation and those in puberty all failed to increase their growth rate. Good results for prepubertal operations to encourage catch up growth were achieved in the Chicago study, where five of seven children improved their height from the third to at least the 10th centile. In this study, however, similar results were achieved in three of six comparable children treated medically and the authors point out that the important determinant of growth is restitution of normal nutrition.<sup>13</sup> Perhaps the present position of treatment of growth retardation in children is best summed up by the same group, who feel that surgery should be reserved for those children with growth failure who fail to respond to a maximal medical and nutritional treatment regime.<sup>14</sup>

#### EFFECT OF ANATOMICAL SITES OF CROHN'S DISEASE ON LONG TERM PROGNOSIS

About equal numbers of ileocolonic, small bowel, and anorectal disease have been reported in the largest series of children studied by Farmer; the frequency and pattern of symptoms was similar to that seen in adults.<sup>15</sup> The extent of small bowel disease, however, in this study was not made

clear. In the present results from Birmingham and in previous reports from Yale, the incidence of diffuse small bowel involvement was noticeably more common than in comparable adult series.<sup>3 15</sup> Nine children (13%) in the Birmingham study had this distribution of disease at presentation and a further five developed diffuse involvement of small bowel during follow up. This compares with only 4% of an adult series reported from Birmingham having diffuse small bowel disease, while none in this group of patients developed it during follow up.<sup>7</sup>

Except for relatively minor operations to relieve small areas of severe stenosis, children with diffuse small bowel disease have to be treated medically and fare poorly, with five out of the total nine deaths in the Birmingham study<sup>3</sup> occurring in this group of patients. Likewise growth retardation which failed to respond to treatment was particularly likely to occur.

#### TREATMENT OF CROHN'S DISEASE IN CHILDHOOD AND THE EFFECT ON LONG TERM PROGNOSIS

The management in this group of patients, consisting of corticosteroids and/or operation, may affect the prognosis. Controversy concerning the choice of these two treatments is highlighted by a comparison of the Yale series,<sup>14</sup> where virtually all the patients were treated with steroids, with the Birmingham study, where steroids were used sparingly for non-obstructive ileal disease, Crohn's colitis, or diffuse small bowel disease. In the Birmingham study only 16 of 38 children with ileocolonic disease received steroids at any stage during follow up, but a more aggressive surgical approach was adopted. Thirty six of those 38 children had at least one operation and the cumulative reoperation rates after 10 years was 50%, including a number of relatively minor revisions of anastomotic areas. This compares with 25 out of the 45 patients with ileocolic disease in the Yale study who underwent surgery.<sup>14</sup>

Most gastroenterologists believe that a more conservative surgical approach with preservation of bowel has resulted in a considerable improvement in the management of Crohn's disease. It is salutary to remember, however, that the Birmingham group, much influenced by Cooke, have produced equally good long term results in both adults and children with a higher operative rate and a much more conservative approach to the use of steroids.

It seems that Crohn's disease in children should not be artificially separated from the spectrum of adult disease, as the symptoms, course, and prognosis are similar. The incidence of diffuse small bowel disease with a relatively gloomy prognosis, however, may be higher in children and growth retardation is a particular problem. The treatment of growth retardation should be directed towards optimum nutrition in the prepubertal child and this is sometimes achieved only with the aid of surgery. Yet again the present results from the Birmingham study<sup>3</sup> indicate that an optimistic approach should be adopted with the parents and the child, as most children go on to lead productive and happy lives.

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